Correlation of Prenatal MRI and Autopsy Findings in the Diagnosis of Vein of Galen Arteriovenous Malformation

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Key words: vein of Galen arteriovenous malformation, prenatal MRI, fetal malformation, transcranial Doppler sonography, obstetric ultrasound scanning

Summary

We report the case of an arteriovenous malformation (AVM) of the vein of Galen diagnosed in utero *at the 34th week of gestation by ultrasound and MRI. Following interdisciplinary advice, the family decided to terminate the pregnancy. This was carried out in the 36th week of gestation. Mascroscopic study of the fetus confirmed the* in utero *diagnosis: considerable dilatation of the cerebral arteries, the Galen vein and the anteroinferior sinuses. Based on the correlation of pre and post natal examinations, this study highlights the accuracy of* in utero *MRI, coupled with ultrasound scanning in the diagnosis of an AVM of the Galen vein. It provides specific information on the degree of cerebral impairment. MRI* in utero *helps in decision-making for pregnancy termination.*

Introduction

Vein of Galen arteriovenous malformation can be discovered by prenatal sonography. MRI *in utero* confirms the diagnosis, indicates the degree of veinous dilatation and the repercussions on the brain, through ventricular dilatation and possible intraparenchymal hypersignal. When all this information is taken into account, adequate treatment is implemented.

Case Report

An expectant mother aged thirty-three, in her 34th week of pregnancy was referred to us following the diagnosis of an aneurysmal malformation of the Galen vein in the fetus, during the last trimester obstetric ultrasound scanning. The results of this examination revealed a biparietal diameter of 92 mm and a cranial perimeter of 317 mm. The Galen vein dilatation measured 27 x 20 mm whilst the median vein of the prosencephalon was 7mm of diameter. Transcranial Doppler sonography displayed blood vessel dilatation of the Circle of Willis; the resistance index on the middle cerebral artery was evaluated at 0.4.

Echocardiography revealed cardiomegaly (cardiothoracic index of 0.78) mainly of the right cavities, associated with a minor pericardial effusion, a slight tricuspid insufficiency; however, kinetics was not affected; heart rate was normal: 130 beats/minute.

Two MRI examinations were performed on a 1 Tesla machine at a fortnight's interval, following the same protocol that included a T1-weighted sagittal plane, and three T2-weighted planes.

The first prenatal MRI examination (13th June 2000) confirmed the diagnosis of an arteriovenous malformation of the Galen vein associated with a dilatation of the venous branch of this fistula (27 mm for the antero-posterior axis and 19 mm for the tranverse diameter); an 8 mm dilatation of the median vein of the prosencephalon, dilatation of the superior sagittal sinus and of the torcular. Biparietal diameter (BPD) was at 92 mm. Dilatation occurred in the lateral ventricles and the subarachnoid peri-cerebral spaces. Signal anomalies were depicted in the periventricular white matter.

A second obstetric ultrasound examination

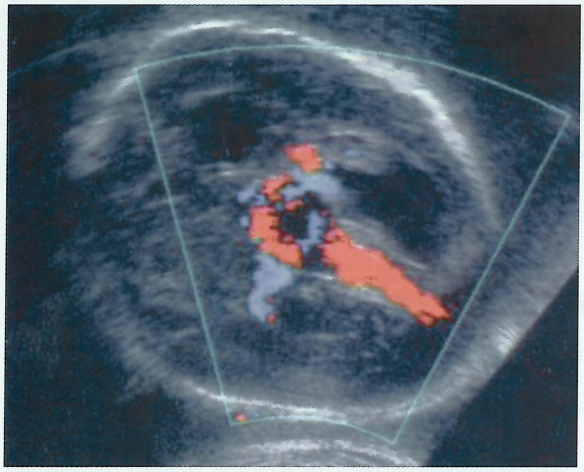


Figure 1 Doppler examination in utero revealed the dilatation of the Galen vein and the increased blood flow within the venous pouch and the falcine sinus.

was carried out on the 22nd June 2000. It highlighted an accentuation of the ventricular dilatation and the biparietal diameter (96mm); cerebral atrophy was also noted in the parietooccipital region and a slight increase was perceived in the hepatomegaly and the pericardial effusion.

A second MRI examination (26th June 2000) delineated aggravation of the Galen vein which measured 34 mm for the anterio-posterior axis and 21 mm for the transverse diameter, and a rise in the BPD to 97 mm and an increase in brain atrophy.

Following the meeting of a multidisciplinary council, the diagnosis and poor prognosis with regards to cerebral and cardiovascular decompensation at birth was announced to the family.

They chose to terminate the pregnancy. This was done on the 36th week of gestation. Arterial opacification, macroscopic and cerebral microscopic studies were achieved. Opacification of the cerebral arteries through the umbilical artery depicted significant dilatation of the cerebral arteries, the Galen vein and the anteroinferior sinuses. Macroscopic findings showed a dilated superficial vein and a ventricular dilatation whilst cardiac microscopic findings depicted diffuse cardiomyopathic hyperplasia.

Discussion

Arteriovenous malformation of the vein of Galen is a rare congenital embryopathy diagnosed mostly during the neonatal period or in childhood. Severe conditions are associated with a life-threatening congestive cardiac failure at birth. The mural-type malformation and the choroidal type lesion consist of a direct shunt between the feeding arteries and the dilated Galen vein draining into an abnormal falcine sinus [1]. This fistula occurs in early embryogenesis, probably before the 11th week, as the falcine sinus observed corresponds to the persistence of the median vein of the prosencephalon; this precursor normally disappears at the 50 mm stage.

Only 20 prenatal cases were described in the literature [2]. This vascular malformation often remains unidentified during the prenatal period but with the improvement in obstetric ultrasound scanning and especially MRI techniques, more prenatal cases will be diagnosed [3]. MRI is crucial because most of the prenatal cases diagnosed and reported in the literature, involve cardiac abnormalities which recurrently reflect an aggressive lesion that brings about neonatal cerebral ischemia and encephalomalacia [4]. In the Necker children's hospital experience, prenatal diagnosis was made in one out of fourteen patients [5]. This is mandatory in order to evaluate the severity of the shunt and the damage that the brain incurred [2]. This diagnosis gives rise to the following questions: how is the cerebral prognosis established? Can it be correlated with brain status on MRI in order to put forward sound arguments for pursuing pregnancy?

We observed a direct fistula between the choroidals and anterior cerebral arteries and the venous pouch but there was no intermediate nidus; this corresponded to a mural-type malformation which does not involve the intermediate entanglement of vessels. The straight sinus was absent whereas the falcine sinus persisted. Both MRI revealed the existence of a venous dilatation which indicated the presence of an arteriovenous shunt and an arterial steal from the brain to the fistula [1]. Ventricular enlargement and the increased pericerebral spaces is synonymous with cerebrospinal fluid disorders. The hypersignal seen on periventricular white matter is most likely of the same origin. The correlation between MRI hypersignal, the gravity of the arteriovenous shunt and the increase in venous pressure is yet to be established. It would be interesting to establish a comparaison between this aspect and the nor-

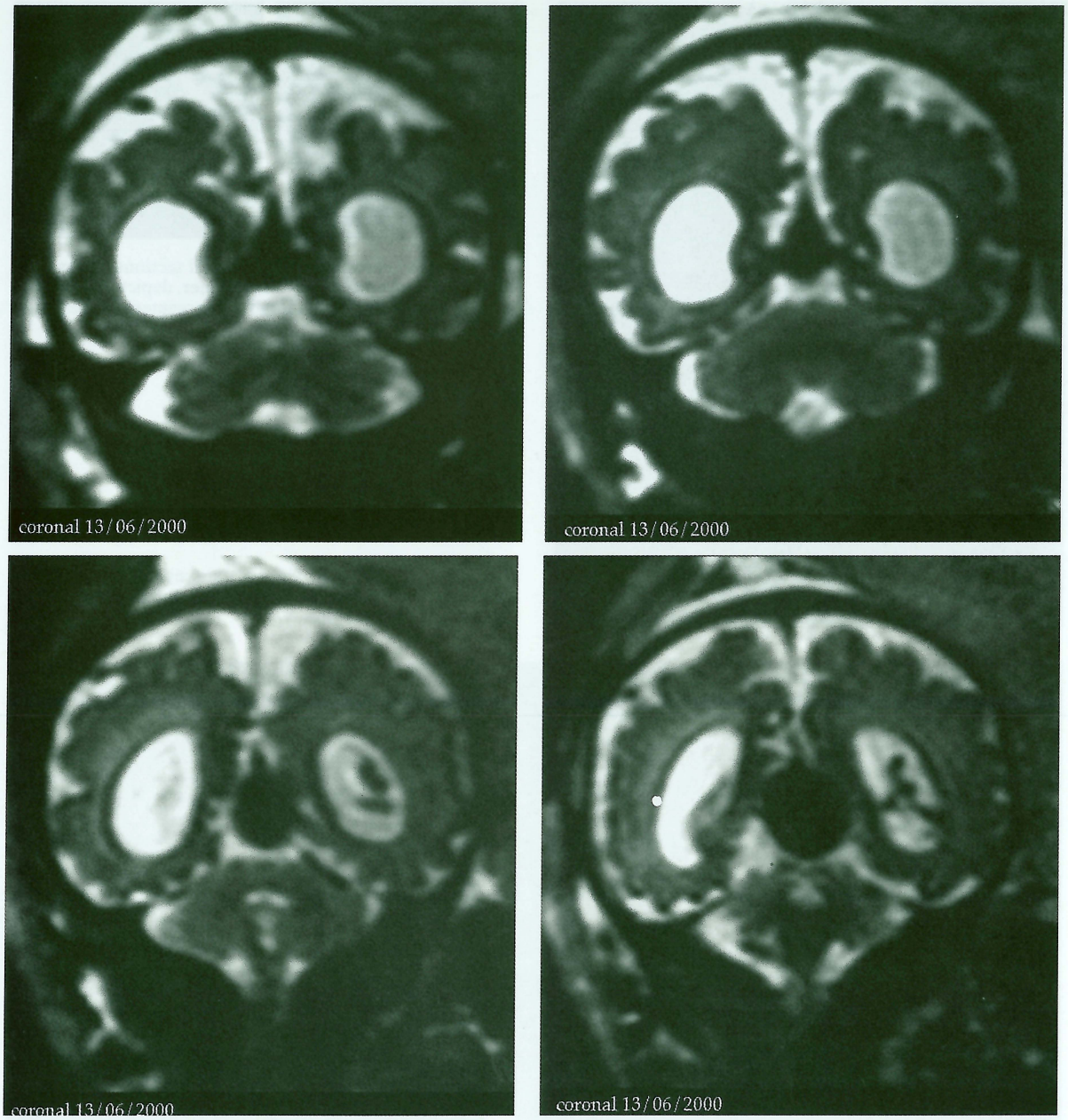
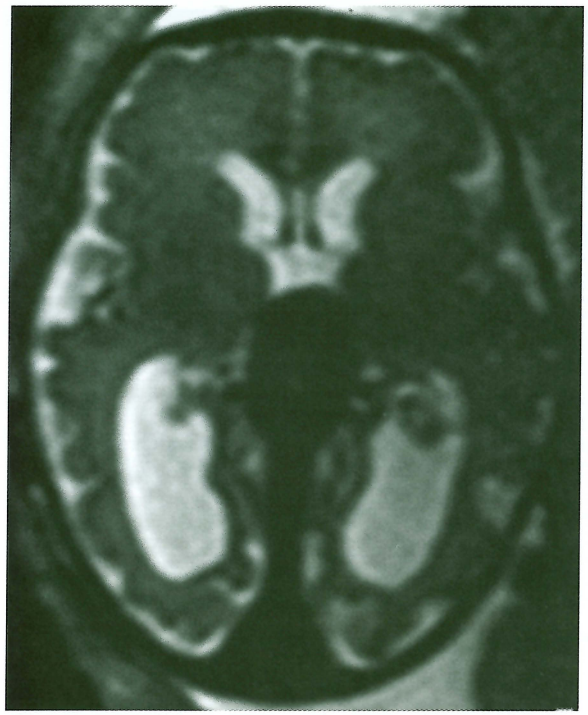


Figure 2 The first MRI (13-6-2000) displayed cerebral atrophy, dilatation of the Galen vein and the lateral ventricles, and white matter hypersignal in the periventricular area mostly on the right.

mal maturation of the brain [2,6,7]. Prenatal brain MRI showed diffuse, high signal intensity from week 31 to the term of pregnancy; this may possibly extend at week 35 to the posterior limbs of the internal capsules, the optic radiations, the midbrain and the central areas of the brain [6]. The considerable increase in cellularity and the evolving processes of myelination are the factors responsible for this hypersignal [6]. The characteristics delineated in those normal cases were significantly different from those depicted asymmetrically in the periventricular areas of both MRI in our case report. No ischemic abnormality was revealed by the histologic examination performed in the same area. However, this microscopic study does not possess the capacity to depict reversible hydrodynamic dysfunctions. MRI is undoubtedly the



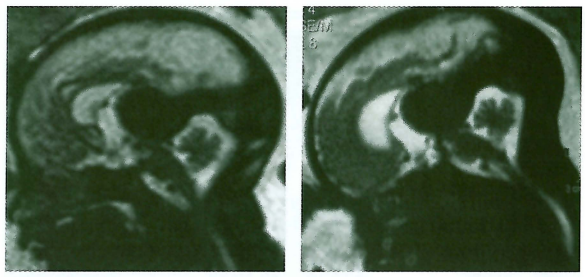


Figure 4 Comparison between sagittal sections on first and second MRI performed a fortnight later, depicted increased cerebral atrophy and considerable enlargement of the Galen vein.

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Figure 3 The first MRI on the transverse section showed the dilatation of the venous side of the shunt and the dilatation of the falcorial sinus.

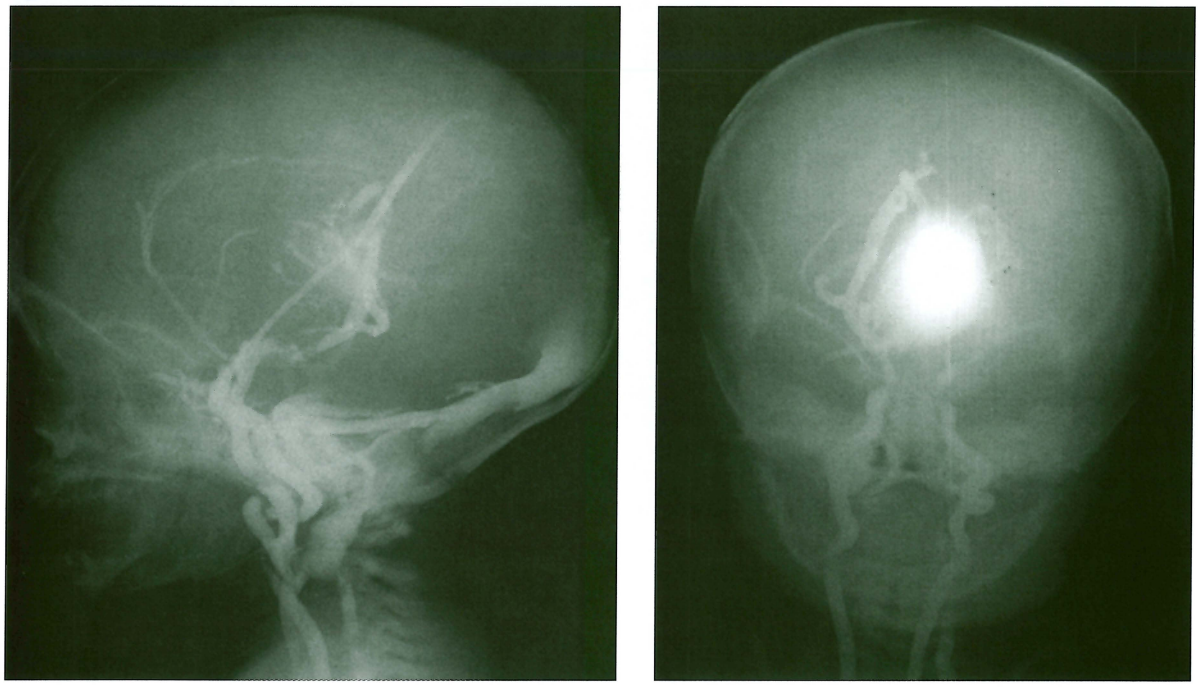

Figure 5 Arterial opacification after abortion through an umbilical arterial punction, depicted the dilatation of the afferent arteries and the venous pouch.

best tool in delineating the morphologic pattern of the malformations, but difficulties occur when it comes to establishing a prognosis and evaluating cerebral hydrodynamic disorders.

The general prognosis to which our multidisciplinary team concluded was bleak with a high risk of morbidity and mortality at birth. This prognosis was based on the increase in the fe-

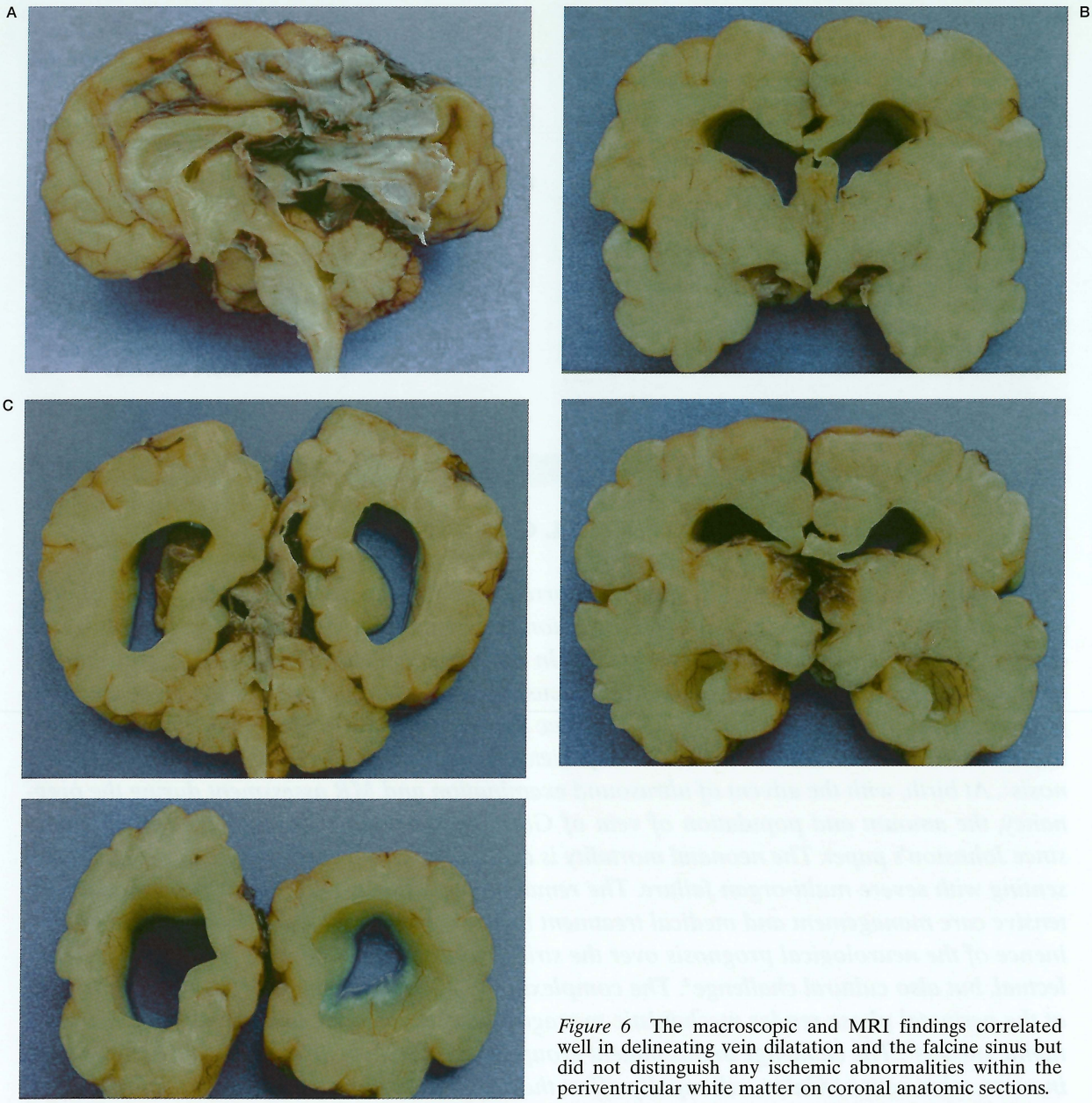

Figure 6 The macroscopic and MRI findings correlated well in delineating vein dilatation and the falcine sinus but did not distinguish any ischemic abnormalities within the periventricular white matter on coronal anatomic sections.

tus's head circumference between the two prenatal MRI examinations, with a cerebral atrophy and a hypersignal of the periventricular white matter.

Spontaneous mortality recorded for neonates with congestive heart failure was 100% in the Johnston review[8], 95% after they had undergone medical treatment, 82% after surgery, and about 75% after emergency endovascular treatment.

In our case report, the mural-type malformation may be reassuring during the neonate period because this pattern of Galenic vascular malformation often becomes symptomatic in infancy when accompanied by neurological symptoms. The cardiomegaly depicted on echocardiography was well-tolerated with a heart rate of 120 beats/minute. It may remain asymptomatic during the first year of life. The macrocephaly already diagnosed *in utero* and its rapid growth is often revealing during childhood when this mural-type malformation is associated with neurological symptoms and a poor prognosis.

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EDITORIAL COMMENT

This article provides interesting anatomic correlations with an antenatally diagnosed of vein of Galen arteriovenous malformation. Discussion concerning the neurological prognosis of the child after birth is of paramount importance. In our experience in Bicêtre, the diagnosis of in utero cardiac failure (fetus pulse above 200, tricuspid insufficiency, extra systole) is the most pejorative sign of rapid brain damage. The cardiac hypertrophy and cerebral ventricular enlargement when associated with an increased bi-parietal diameter does not seem to carry a bad prognosis[1]. At birth, with the advent of ultrasound examination and MR assessment during the pregnancy, the amount and population of vein of Galen malformation has increased significantly since Johnston's paper. The neonatal mortality is around 35% and corresponds to neonates presenting with severe multi-organ failure. The remaining two thirds will benefit from modern intensive care management and medical treatment before transarterial embolization. The pre-eminence of the neurological prognosis over the strict survival of the neonate is a technical, intellectual, but also cultural challenge[2]. The complexity of the anatomy and physiology of the fetus at the perinatal phase render mechanistic management gross despite some immediate hemodynamic success[3]. The challenge in this patient group remains our capacity to predict early enough in utero, what vein of Galen will rapidly affect the developing brain. It is to be remembered that most of these lesions diagnosed in utero today will not lead to severe cardiac failure at birth, and that some of the severe neonatal forms are not seen on good antenatal ultrasound examinations.

P. Lasjaunias

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